Acute hemorrhagic edema of infancy

Pei-Rong Gao MD, Hsi Yen MD MPH, Wei-Ti Chen MD MS

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A 6-month-old, full-term infant girl was brought to the emergency department after 3 days of having an asymptomatic, large, purple-red bruiselike rash (Figure 1). The rash first appeared on the patient's thighs bilaterally before spreading rapidly to her face, abdomen and lower legs. She had an episode of acute viral gastroenteritis that had resolved 1 week earlier, but she was otherwise healthy. An examination of her skin showed medallionlike, well-defined ecchymoses (2–3 cm in diameter) with surrounding erythematous edematous plaques affecting the face, periumbilical area and extremities (Figure 1); the lesions were warm to the touch. The patient had edema but no fever.

Results from laboratory tests showed hypocomplementemia. A biopsy of the patient's skin found perivascular infiltrate with numerous neutrophils, some eosinophils, nuclear dust, abundant extravasated erythrocytes and hemorrhage (Appendix 1, available at www.cmaj.ca/lookup/doi/10.1503/cmaj.200418/tab-related-content). Direct immunofluorescence was negative for immunoglobulin deposition. The pathology was compatible with leukocytoclastic vasculitis. We diagnosed acute hemorrhagic edema of infancy. The patient's skin lesions improved spontaneously without specific treatment over a 2-week period.

Acute hemorrhagic edema of infancy is an uncommon disorder with a reported incidence of 0.7 cases per 1000 admissions in one case series, mainly affecting infants and children under 2 years of age.[1,2] Although the cause is unclear, the disease may be preceded by infections (e.g., upper respiratory infection or gastroenteritis), medications (e.g., acetaminophen or penicillin) and immunization; an immune complex–mediated pathogenesis has been hypothesized.[1–3] The classic clinical triad includes fever, edema and round, palpable, purpuric plaques on the face and extremities.[2] Leukocytosis and hypocomplementemia may be found on examination of the blood, with hematuria and proteinuria noted on urinalysis.[1,4] A skin biopsy showing leukocytoclastic vasculitis is diagnostic.[1–3]

The differential diagnosis of acute hemorrhagic edema of infancy includes Henoch–Schönlein purpura, urticaria and erythema multiforme.[1–3] Acute hemorrhagic edema of infancy may be mistaken for lesions associated with child abuse; thorough screening with prompt reporting as per relevant protocols must be conducted if there is any concern.[3] The disease generally resolves without treatment within 1–3 weeks.[1–3]

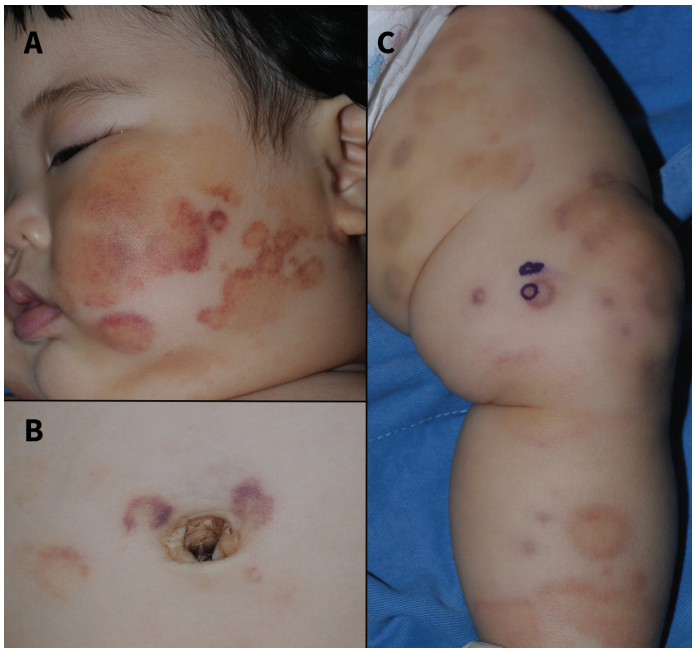

Figure 1: A 6-month-old girl with acute hemorrhagic edema of infancy who presented with medallionlike, well-defined ecchymoses with surrounding erythematous edematous plaques on (A) the face, (B) periumbilical area and (C) left leg. The circle of purple ink on the patient's left thigh shows the site of biopsy.

References

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Affiliations: Department of Dermatology (Gao, Yen, Chen) and Drug Hypersensitivity Clinical and Research Center (Chen), Chang Gung Memorial Hospital, Linkou, Taiwan; College of Medicine (Gao, Yen, Chen), Chang Gung University, Taoyuan, Taiwan; Graduate Institute of Immunology (Chen), College of Medicine, National Taiwan University, Taipei, Taiwan

Correspondence to: Wei-Ti Chen, greatedisonchen@gmail.com